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Case reports

Management of a patient with Wilms's tumour extending into the right heart chambers: a case report and a review of other published reports

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SUMMARY Wilms's tumours that extend by direct intravascular spread into the right side of the heart are rare. A case of such a tumour was diagnosed by ultrasound. A one stage resection was performed on cardiopulmonary bypass and with profound hypothermic circulatory arrest. It was followed by adjunctive chemotherapy and radiotherapy. The child was alive and tumour free 18 months later. A review of 17 other similar cases indicated that multimodal treatment is justified in patients with extensive intravascular spread of Wilms's tumours.

There have been only 17 cases of right atrial extension of Wilms's tumour reported in English this century.[1-14] Such cases used to be regarded as inoperable. Advances in surgical techniques and adjunctive treatment have made surgical excision possible and worth while even when the disease is extensive. We report the management of a case of this type of Wilms's tumour which we believe to be the first reported from Britain, and we review earlier cases.

Case report

A five and a half year old boy was admitted with a history of lethargy and diarrhoea for three weeks and generalised abdominal pain and anorexia for one week.

On examination he was unwell with a tachycardia, tachypnoea, and leg oedema. His blood pressure was 110/70 mm Hg but the jugular venous pressure was raised with a prominent "a" wave. A systolic ejection murmur was audible at the left sternal edge. There were signs of a right pleural effusion. The abdomen was distended with considerable hepatomegaly, a mass in the left upper quadrant, and ascites.

Investigations showed a haemoglobin concentration of 142 g/l, white cell count of 15·8 × 10^9/l, and platelet count of 40 × 10^9/l. Coagulation tests showed an International Normalised Ratio of 2·1. The serum activity of alanine aminotransferase was raised (1266 IU/l). Bilirubin and renal function were normal.

Chest *x* ray confirmed a right pleural effusion and enlarged cardiac silhouette. Ultrasound scan confirmed the presence of ascites and a pleural effusion, and showed a grossly enlarged left kidney with distorted calyces. The inferior vena cava was occluded by tumour or thrombus that extended into the right atrium and prolapsed into the right ventricle with each cardiac cycle. A diagnosis of Wilms's tumour originating in the left kidney and extending to the inferior vena cava and right heart was made on the basis of this investigation.

Computed tomography showed that the renal mass originated in the upper pole and showed dilatation of the inferior vena cava. It did not detect extension of tumour into the right atrium. The lung fields were clear. Venography showed obstruction of the inferior vena cava and iliac veins above the internal iliac vessels with drainage via anastomotic channels to the vertebral venous system.

Real time cross sectional echocardiography confirmed the intracardiac tumour and suggested that it was mobile (figure). A skeletal survey was normal.

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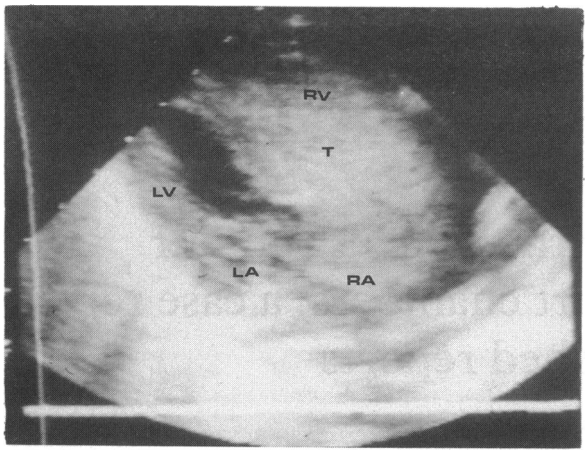

Figure *Echocardiogram (apical four chamber view) showing tumour within right atrium prolapsed through to the right ventricle. T, tumour; RV, RA, LV, LA, heart chambers.*

OPERATION

A cardiac team removed the intravascular tumour and an abdominal team performed the radical nephrectomy. A routine cardiac surgical anaesthetic was given. Intravascular lines were placed to monitor arterial blood pressure, pressure in the inferior and superior caval veins, and to administer fluid. A urinary catheter was inserted. Temperature probes were placed in the nasopharynx, oesophagus, and on the tympanic membrane.

A median sternotomy was performed, the presence of a large tumour mass in the right atrium was confirmed, and the heart was prepared for cardiopulmonary bypass. The incision was then extended into a full midline laparotomy and the left renal tumour was mobilised by the second team of surgeons. The inferior vena cava, left renal vein, adrenal vein, and testicular vein all had clot and tumour extending into them. The liver was grossly congested but showed no evidence of metastases. The testicular vessels were ligated distal to any macroscopic tumour and the ureter was divided at the pelvic brim.

During the final part of the dissection haemodynamic instability developed. The patient was therefore given heparin and cardiopulmonary bypass was established with one right atrial venous cannula and an ascending aortic return. While the patient was cooled to 15°C the radical left nephrectomy was completed. The specimen weighed 260 g. The aorta was cross clamped, the circulation was arrested, and the patient was exsanguinated. The right atrium and infrahepatic vena cava were opened. The intracardiac tumour was mobile but the intracaval portion was adherent and required careful instrumentation from above and below to free it. Fogarty extraction of distal venous thrombosis to the iliac veins was performed and the right renal vein and both pulmonary arteries were checked for clearance. The inferior vena cava and right atrium were repaired and cardiopulmonary bypass was re-established by bicaval cannulation after 37 minutes of circulatory arrest. The patient was rewarmed and came off bypass without difficulty. The chest and abdomen were closed routinely with drainage. There was no difficulty in obtaining haemostasis after reversal of the heparinisation with protamine sulphate.

The patient made an excellent postoperative recovery. Histopathology confirmed the diagnosis of Wilms's tumour of the favourable histological type. Postoperative radiotherapy was given as well as one year of chemotherapy. The patient was alive, well, and disease free 18 months after operation.

Discussion

Direct extension of Wilms's tumour into the right heart cavities is rare. In one study it occurred in 0·7% of cases of Wilms's tumour.[15] The table summarises the clinical features of all 17 reported cases.

Accurate preoperative diagnosis of the extent of the tumour is required to plan the management of each case. Assessment of the inferior vena cava is important in all cases of renal tumour, and before resection of hepatic tumours also, to minimise the risk of tumour embolism at operation and allow for complete resection. The upper limit of intravascular spread needs to be defined. If the caval veins are involved the possibility of extension of the tumour to the heart must be investigated. This avoids the risk that extension to the heart will be recognised only when abdominal surgery is under way and when appropriate facilities for cardiothoracic operation may not be available. Similarly, it is important to assess the inferior vena cava for distal extension in cases of suspected right atrial tumour.[7]

In nine of the 17 reported cases an accurate diagnosis was not made before the initial operation was planned (table). One patient presented with an abdominal Wilms's tumour and the inferior vena cava was not investigated before operation.[4] The inferior vena cava was known to be affected in two cases but no further elective investigations were undertaken to define the upper limit of intravascular extension.[3 8] In the other eight cases, and in the case reported here, the extent of the tumour was recognised before operation. Angiography has been recommended for assessment of the intravascular spread of tumours,[8 9 14] but this invasive procedure risks precipitating tumour embolism or cardiovascular decompensation in patients whose condition is un-

stable because the heart is affected.[10] Newer imaging techniques, particularly ultrasound, are useful for non-invasive investigation.[5 6 11 13 15] We support the use of ultrasonography, which gave better results than computed tomography in our patient.

Several surgical options are available (table). Although staged resection has been performed,[3 10] a one stage procedure is more desirable. If the tumour has extended into the right heart, cardiopulmonary bypass is usually required. We used cardiopulmonary bypass with profound hypothermia and circulatory arrest. A median sternotomy and midline laparotomy gave good access for both surgical teams, and avoids the risk of precipitating acute right heart obstruction when a patient with a mobile atrial tumour extension is turned.[4 16] We performed the sternotomy first to allow rapid institution of cardiopulmonary bypass in the event of haemodynamic instability. Although the whole procedure could have been performed on bypass, we elected to do as much of the dissection as possible before heparinisation to avoid the risk of excessive bleeding. Circulatory arrest allowed controlled, accurate excision of the atrial and caval tumour in a bloodless field, and reduced the risk of tumour embolism. This phase of the resection was completed well within the safe period of circulatory arrest (45–60 minutes at 15 °C). Cardiopulmonary bypass[13 14] and circulatory arrest[3 10] have been used successfully before, but ours is the first reported case in which a planned one stage resection was performed on bypass, profound hypothermia, and circulatory arrest. This technique is similar to that reported in adults with renal cell carcinoma.[17 18]

The prognosis of this type of Wilms's tumour is dependent on accurate preoperative assessment. Only two of the nine patients with an accurate preoperative diagnosis (including our case) have died[5 9] and one of these had unresectable primary disease.[5] The other seven were alive and well between eight months and five years after operation.[10–14] Of the nine with inadequate preoperative assessment, only one has survived for more than six months (three years six months).[3] Another review reported an increased complication rate when preoperative diagnosis was not accurate. This review included some of

Table *Summary of the 17 earlier cases of Wilms's tumour extending into the right atrium*

Ref No	*Authors (date)*	*Method of diagnosis*	*Operation*	*Adjuvant treatment*	*Outcome*
1	Nadas and Ellison (1968) (3 cases)	Necropsy	—	—	—
2	Anselmi *et al* (1970)	Necropsy	—	—	—
3	Murphy *et al* (1973)	Operation at time of nephrectomy	CPB and circulatory arrest the next day	Chemotherapy and irradiation	A and W 3 y and 6 mnth
4	Utley *et al* (1973)	Operation at time of nephrectomy	CPB and circulatory arrest after abdominal closure	Chemotherapy and irradiation	Died 3 mnth
5	Farooki *et al* (1975)	Echo and cardiac catheter	Unresectable primary	Chemotherapy and irradiation	Died
6	Farooki *et al* (1976)	Echo 6 mnth after previous nephrectomy	1 stage resection with CPB	None specified	Died 6 mnths (Ref 11)
7	Aytac *et al* (1976)	Cardiac catheter diagnosis of atrial tumour. Extension from below diaphragm only recognised at operation. Wilms's tumour diagnosed at necropsy	Atrial clearance only with CPB	—	Died
8	Schullinger *et al* (1977)	Emergency cardiac catheterisation after elective operation had been planned	1 stage resection with CPB	—	Died at operation
9	Vaughan *et al* (1977)	Cardiac catheterisation	1 stage resection with CPB via T A incision	Chemotherapy and irradiation	Died 6 mnth
10	Theman *et al* (1978)	Cardiac catheterisation and CT scan	2 stages with CPB and circulatory arrest, and "interval nephrectomy" after 3 mnth	Chemotherapy and irradiation	A and W 19 mnth
11	Slovis *et al* (1978)	Cardiac catheterisation and CT scan	No treatment described	?	Alive 8 mnth
12	Kolmannskog *et al* (1979)	Cardiac catheterisation	1 stage resection with no CPB	Chemotherapy only	A and W 15 mnth
13	Luck *et al* (1982) (2 cases)	Echo and ultrasound	1 stage resection with CPB	Chemotherapy and irradiation	A and W 18 mnth; A and W 20 mnth
14	Schraut and Chilcote (1985)	Cardiac catheterisation	1 stage resection with CPB	Chemotherapy and irradiation	A and W 5 y

CPB, cardiopulmonary bypass; T A, thoraco-abdominal; A and W, alive, well, disease free; CT, computed tomography.

the reported cases reviewed in the present article.[15]

Intravascular extension of Wilms's tumour into the right side of the heart is rare. With modern imaging techniques, particularly ultrasound, it should be possible, however, to establish a diagnosis before operation. A planned one stage resection is then feasible after a midline sternotomy and laparotomy. Cardiopulmonary bypass, with or without hypothermic circulatory arrest, is needed for excision of the intravascular tumour. Complete surgical resection followed by adjunctive chemotherapy and radiotherapy may give long term, tumour free survival.

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